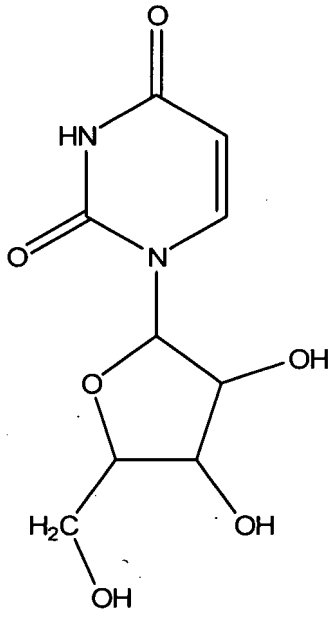
IN THE CLAIMS

Please amend claims as shown below. Please add new claims 111-180. The following is the listing of the claims replacing all previous claims.

1-66. (Canceled).

67. (Currently amended) A method for the treatment of a mitochondrial disorder comprising administering to a subject having or at risk of having such disorder an effective amount of L isomer or D isomer of a keto tautomer or a enol tautomer of a compound, ~~said~~ the keto tautomer having the Formula I, and ~~said~~ the enol tautomer having the Formula IA:



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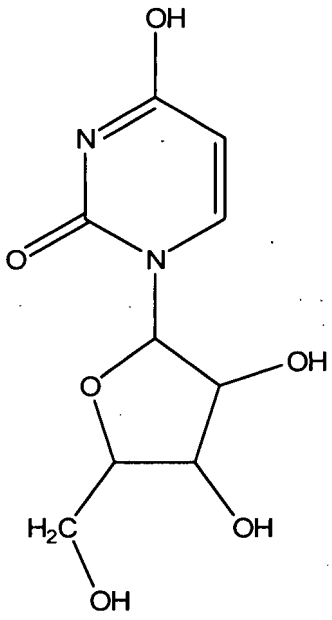


IA

wherein the mitochondrial disorder is selected from a group consisting of mitochondrial renal tubular acidosis, multiple mitochondrial deletion syndrome, Leigh syndrome, lactic acidemia, 3-hydroxybutyric acidemia, encephalomyopathy, 1+proteinuria, pyruvate

dehydrogenase deficiency, complex I deficiency, complex IV deficiency, aminoaciduria, hydroxyprolinuria, and MARIAHS syndrome, and wherein the compound is selected from uridine and 1-β-D-ribofuranosyluracil.

68-69. (Canceled).

70. (Previously presented) The method according to claim 67, wherein the mitochondrial disorder is a primary disorder comprising at least one mutation in mitochondrial or nuclear DNA.

71-72. (Canceled).

73. (Currently amended) The method according to claim 67, wherein ~~said~~ the mitochondrial disorder is a secondary disorder caused by acquired somatic mutations, physiologic effects of drugs, viruses, or environmental toxins that inhibit mitochondrial function.

74. (Previously presented) The method according to claim 67, wherein the mitochondrial disorder is a deficiency of cardiolipin.

75. (Previously presented) The method according to claim 67, wherein the mitochondrial disorder comprises a deficiency in a pyrimidine synthetic pathway.

76. (Previously presented) The method according to claim 75, wherein the deficiency in a pyrimidine synthetic pathway is the deficiency in the uridine synthetic pathway.

77. (Previously presented) The method according to claim 75, wherein the deficiency comprises reduced expression and/or activity of an enzyme in the pyrimidine synthetic pathway.

78. (Previously presented) The method according to claim 77, wherein the enzyme is selected from the group consisting of dihydroorotate dehydrogenase (DHOD) and uridine monophosphate synthetase (UMPS).

79. (Previously presented) The method according to claim 67, wherein the mitochondrial disorder results in lower than normal uridine levels.

80. (Previously presented) The method according to claim 67, wherein the mitochondrial disorder is the result of prior or concurrent administration of a pharmaceutical agent.

81. (Previously presented) The method according to claim 80, wherein the pharmaceutical agent is a reverse transcriptase inhibitor, a protease inhibitor or an inhibitor of DHOD.

82-83. (Canceled).

84. (Previously presented) The method according to claim 81, wherein the DHOD inhibitor is Leflunomide or Brequinar.

85. (Previously presented) The method according to claim 67, further comprising the administration of one or more co-factors, vitamins, or mixtures of two or more thereof.

86. (Previously presented) The method according to claim 85, wherein the co-factor is one or both of Coenzyme Q10 or calcium or magnesium pyruvate.

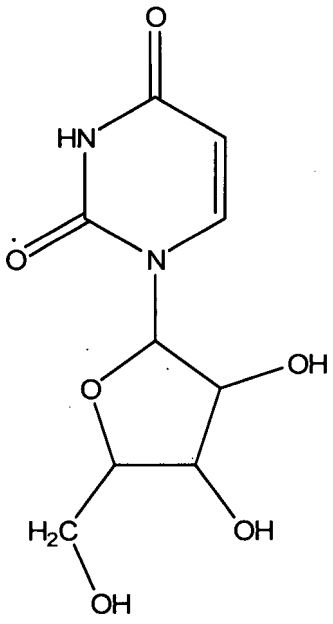
87. (Previously presented) The method according to claim 85, wherein the vitamin is selected from the group consisting of thiamine (B1), riboflavin (B2), niacin (B3), pyridoxine (B6), folate, cyanocobalamine (B12), biotin, α-lipoic acid, and pantothenic acid.

88. (Currently amended) The method according to claim 67, wherein the compound of Formula (I) or Formula (IA) is administered in a daily dosage in the range of about 0.5 g/m^2 to 20 g/m^2.

89. (Currently amended) The method according to claim 67, wherein the compound of Formula (I) or Formula (IA) is administered in a daily dosage in the range of about 2 g/m^2 to 10 g/m^2.

90. (Currently amended) The method according to claim 67, wherein the compound of Formula (I) or Formula (IA) is administered in a daily dosage of about 6.0 g/m^2.

91. (Currently amended) A method for reducing or eliminating one or more symptoms associated with a mitochondrial disorder comprising administering to a subject in need thereof an effective amount of L isomer or D isomer of a keto tautomer or a enol tautomer of a compound, ~~said~~ the keto tautomer having the Formula I, and ~~said~~ the enol tautomer having the Formula IA:



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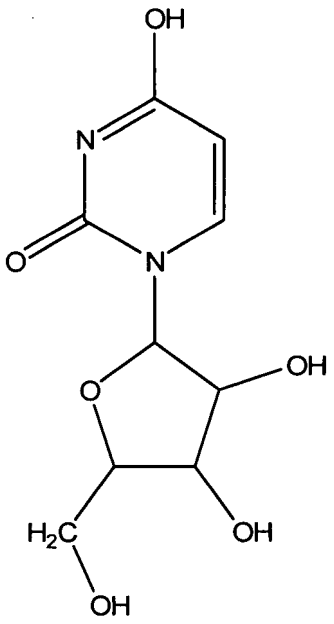


IA

wherein the mitochondrial disorder is selected from a group consisting of mitochondrial renal tubular acidosis, multiple mitochondrial deletion syndrome, Leigh syndrome, lactic acidemia, 3-hydroxybutyric acidemia, encephalomyopathy, 1+proteinuria, pyruvate dehydrogenase deficiency, complex I deficiency, complex IV deficiency, aminoaciduria, hydroxyprolinuria, and MARIAHS syndrome, and wherein the compound is selected from uridine and 1-β-D-ribofuranosyluracil.

92-94. (Canceled).

95. (Previously presented) The method according to claim 67, wherein the mitochondrial disorder is MARIAHS syndrome.

96. (Previously presented) The method according to claim 95, wherein the mitochondrial disorder comprises a deficiency in a pyrimidine synthetic pathway.

97. (Previously presented) The method according to claim 96, wherein the deficiency in a pyrimidine synthetic pathway is the deficiency in the uridine synthetic pathway.

98. (Previously presented) The method according to claim 96, wherein the deficiency comprises reduced expression and/or activity of an enzyme in the pyrimidine synthetic pathway.

99. (Previously presented) The method according to claim 98, wherein the enzyme is selected from the group consisting of dihydroorotate dehydrogenase (DHOD) and uridine monophosphate synthetase (UMPS).

100. (Previously presented) The method according to claim 95, wherein the mitochondrial disorder results in lower than normal uridine levels.

101. (Previously presented) The method according to claim 95, wherein the mitochondrial disorder is the result of prior or concurrent administration of a pharmaceutical agent.

102. (Previously presented) The method according to claim 101, wherein the pharmaceutical agent is a reverse transcriptase inhibitor, a protease inhibitor or an inhibitor of DHOD.

103. (Previously presented) The method according to claim 102, wherein the DHOD inhibitor is Leflunomide or Brequinar.

104. (Previously presented) The method according to claim 95, further comprising the administration of one or more co-factors, vitamins, or mixtures of two or more thereof.

105. (Previously presented) The method according to claim 104, wherein the co-factor is one or both of Coenzyme Q10 or calcium or magnesium pyruvate.

106. (Previously presented) The method according to claim 104, wherein the vitamin is selected from the group consisting of thiamine (B1), riboflavin (B2), niacin (B3), pyridoxine (B6), folate, cyanocobalamine (B12), biotin, α-lipoic acid, and pantothenic acid.

107. (Currently amended) The method according to claim 95, wherein the compound of Formula (I) or Formula (IA) is administered in a daily dosage in the range of about 0.5 g/m^2 to 20 g/m^2.

108. (Currently amended) The method according to claim 95, wherein the compound of Formula (I) or Formula (IA) is administered in a daily dosage in the range of about 2 g/m^2 to 10 g/m^2.

109. (Currently amended) The method according to claim 95, wherein the compound of Formula (I) or Formula (IA) is administered in a daily dosage of about 6.0 g/m^2.

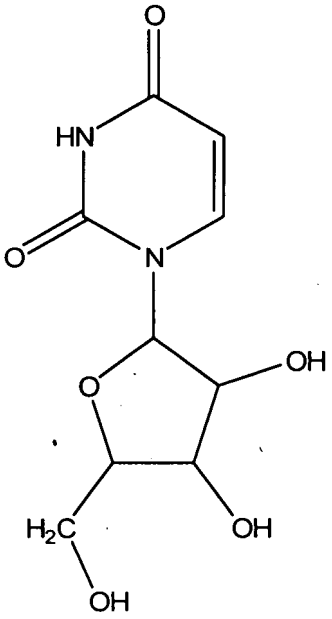
110. (Previously presented) The method according to claim 91, wherein the mitochondrial disorder is MARIAHS syndrome.

111. (New) A method for the treatment of a mitochondrial disorder comprising administering to a subject having or at risk of having such disorder a pharmaceutical composition consisting of:

(a) an effective amount of L isomer or D isomer of a keto tautomer or a enol tautomer of a compound; and

(b) a pharmaceutically acceptable vehicle,

wherein the keto tautomer has the Formula I, and the enol tautomer having the Formula IA:



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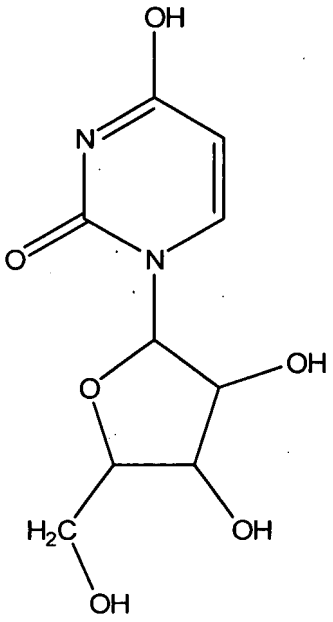


IA

and wherein the mitochondrial disorder is selected from a group consisting of mitochondrial renal tubular acidosis, multiple mitochondrial deletion syndrome, Leigh syndrome, lactic acidemia, 3-hydroxybutyric acidemia, encephalomyopathy, 1+proteinuria, pyruvate dehydrogenase deficiency, complex I deficiency, complex IV deficiency, aminoaciduria, hydroxyprolinuria, and MARIAHS syndrome,

with the further proviso that the compound is selected from uridine and 1-β-D-ribofuranosyluracil.

112. (New) The method according to claim 111, wherein the mitochondrial disorder is a primary disorder comprising at least one mutation in mitochondrial or nuclear DNA.

113. (New) The method according to claim 111, wherein the mitochondrial disorder is a secondary disorder caused by acquired somatic mutations, physiologic effects of drugs, viruses, or environmental toxins that inhibit mitochondrial function.

114. (New) The method according to claim 111, wherein the mitochondrial disorder is a deficiency of cardiolipin.

115. (New) The method according to claim 111, wherein the mitochondrial disorder comprises a deficiency in a pyrimidine synthetic pathway.

116. (New) The method according to claim 115, wherein the deficiency in a pyrimidine synthetic pathway is the deficiency in the uridine synthetic pathway.

117. (New) The method according to claim 115, wherein the deficiency comprises reduced expression and/or activity of an enzyme in the pyrimidine synthetic pathway.

118. (New) The method according to claim 117, wherein the enzyme is selected from the group consisting of dihydroorotate dehydrogenase (DHOD) and uridine monophosphate synthetase (UMPS).

119. (New) The method according to claim 111, wherein the mitochondrial disorder results in lower than normal uridine levels.

120. (New) The method according to claim 111, wherein the mitochondrial disorder is the result of prior or concurrent administration of a pharmaceutical agent.

121. (New) The method according to claim 120, wherein the pharmaceutical agent is a reverse transcriptase inhibitor, a protease inhibitor or an inhibitor of DHOD.

122. (New) The method according to claim 121, wherein the DHOD inhibitor is Leflunomide or Brequinar.

123. (New) The method according to claim 111, further comprising the administration of one or more co-factors, vitamins, or mixtures of two or more thereof.

124. (New) The method according to claim 123, wherein the co-factor is one or both of Coenzyme Q10 or calcium or magnesium pyruvate.

125. (New) The method according to claim 123, wherein the vitamin is selected from the group consisting of thiamine (B1), riboflavin (B2), niacin (B3), pyridoxine (B6), folate, cyanocobalamine (B12), biotin, α-lipoic acid, and pantothenic acid.

126. (New) The method according to claim 111, wherein the pharmaceutical composition is administered in a quantity providing for delivering the compound of Formula (I) or (IA) in a daily dosage in the range of about 0.5 g/m^2 to 20 g/m^2.

127. (New) The method according to claim 111, wherein the pharmaceutical composition is administered in a quantity providing for delivering the compound of Formula (I) or (IA) in a daily dosage in the range of about 2 g/m^2 to 10 g/m^2.

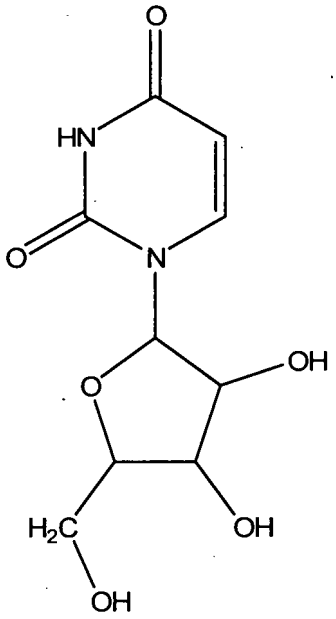
128. (New) The method according to claim 111, wherein the pharmaceutical composition is administered in a quantity providing for delivering the compound of Formula (I) or (IA) in a daily dosage of about 6.0 g/m^2.

129. (New) A method for reducing or eliminating one or more symptoms associated with a mitochondrial disorder comprising administering to a subject having or at risk of having such disorder a pharmaceutical composition consisting of:

(a) an effective amount of L isomer or D isomer of a keto tautomer or a enol tautomer of a compound; and

(b) a pharmaceutically acceptable vehicle,

wherein the keto tautomer has the Formula I, and the enol tautomer has the Formula IA:



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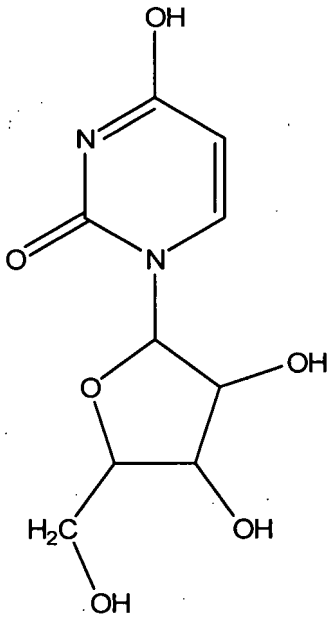

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and wherein the mitochondrial disorder is selected from a group consisting of mitochondrial renal tubular acidosis, multiple mitochondrial deletion syndrome, Leigh syndrome, lactic acidemia, 3-hydroxybutyric acidemia, encephalomyopathy, 1+proteinuria, pyruvate dehydrogenase deficiency, complex I deficiency, complex IV deficiency, aminoaciduria, hydroxyprolinuria, and MARIAHS syndrome,

with the further proviso that the compound is selected from uridine and 1-β-D-ribofuranosyluracil.

130. (New) The method according to claim 111, wherein the mitochondrial disorder is MARIAHS syndrome.

131. (New) The method according to claim 130, wherein the mitochondrial disorder comprises a deficiency in a pyrimidine synthetic pathway.

132. (New) The method according to claim 131, wherein the deficiency in a pyrimidine synthetic pathway is the deficiency in the uridine synthetic pathway.

133. (New) The method according to claim 131, wherein the deficiency comprises reduced expression and/or activity of an enzyme in the pyrimidine synthetic pathway.

134. (New) The method according to claim 133, wherein the enzyme is selected from the group consisting of dihydroorotate dehydrogenase (DHOD) and uridine monophosphate synthetase (UMPS).

135. (New) The method according to claim 130, wherein the mitochondrial disorder results in lower than normal uridine levels.

136. (New) The method according to claim 130, wherein the mitochondrial disorder is the result of prior or concurrent administration of a pharmaceutical agent.

137. (New) The method according to claim 136, wherein the pharmaceutical agent is a reverse transcriptase inhibitor, a protease inhibitor or an inhibitor of DHOD.

138. (New) The method according to claim 137, wherein the DHOD inhibitor is Leflunomide or Brequinar.

139. (New) The method according to claim 130, further comprising the administration of one or more co-factors, vitamins, or mixtures of two or more thereof.

140. (New) The method according to claim 139, wherein the co-factor is one or both of Coenzyme Q10 or calcium or magnesium pyruvate.

141. (New) The method according to claim 139, wherein the vitamin is selected from the group consisting of thiamine (B1), riboflavin (B2), niacin (B3), pyridoxine (B6), folate, cyanocobalamine (B12), biotin, α-lipoic acid, and pantothenic acid.

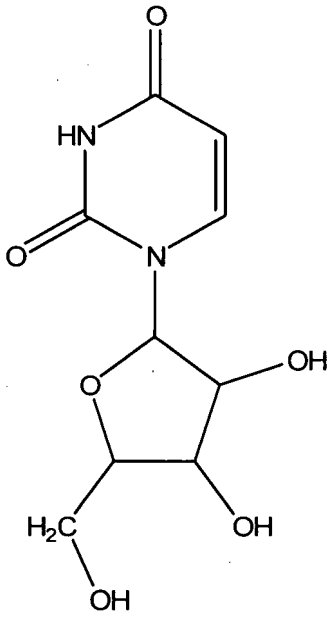
142. (New) The method according to claim 130, wherein the pharmaceutical composition is administered in a quantity providing for delivering the compound of Formula (I) or (IA) in a daily dosage in the range of about 0.5 g/m^2 to 20 g/m^2.

143. (New) The method according to claim 130, wherein the pharmaceutical composition is administered in a quantity providing for delivering the compound of Formula (I) or (IA) in a daily dosage in the range of about 2 g/m^2 to 10 g/m^2.

144. (New) The method according to claim 130, wherein the pharmaceutical composition is administered in a quantity providing for delivering the compound of Formula (I) or (IA) in a daily dosage of about 6.0 g/m^2.

145. (New) The method according to claim 129, wherein the mitochondrial disorder is MARIAHS syndrome.

146. (New) A method for the treatment of a mitochondrial disorder comprising administering to a subject having or at risk of having such disorder an effective amount of L isomer or D isomer of a keto tautomer or a enol tautomer of a compound, the keto tautomer having the Formula I, and the enol tautomer having the Formula IA:



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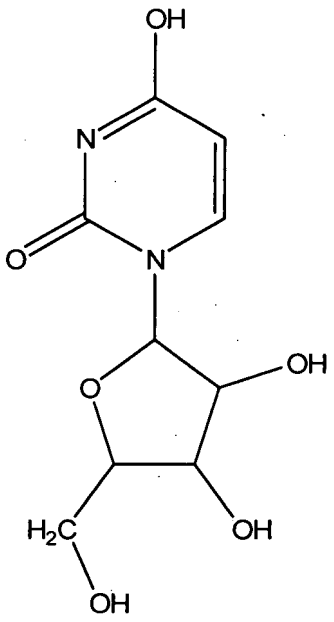


IA

wherein the mitochondrial disorder is selected from a group consisting of mitochondrial renal tubular acidosis, multiple mitochondrial deletion syndrome, Leigh syndrome, lactic acidemia, 3-hydroxybutyric acidemia, 1+proteinuria, pyruvate dehydrogenase deficiency, complex I deficiency, complex IV deficiency, aminoaciduria, hydroxyprolinuria, and MARIAHS syndrome, and wherein the compound is selected from uridine and 1-β-D-ribofuranosyluracil.

147. (New) The method according to claim 146, wherein the mitochondrial disorder is a primary disorder comprising at least one mutation in mitochondrial or nuclear DNA.

148. (New) The method according to claim 146, wherein the mitochondrial disorder is a secondary disorder caused by acquired somatic mutations, physiologic effects of drugs, viruses, or environmental toxins that inhibit mitochondrial function.

149. (New) The method according to claim 146, wherein the mitochondrial disorder is a deficiency of cardiolipin.

150. (New) The method according to claim 146, wherein the mitochondrial disorder comprises a deficiency in a pyrimidine synthetic pathway.

151. (New) The method according to claim 150, wherein the deficiency in a pyrimidine synthetic pathway is the deficiency in the uridine synthetic pathway.

152. (New) The method according to claim 150, wherein the deficiency comprises reduced expression and/or activity of an enzyme in the pyrimidine synthetic pathway.

153. (New) The method according to claim 152, wherein the enzyme is selected from the group consisting of dihydroorotate dehydrogenase (DHOD) and uridine monophosphate synthetase (UMPS).

154. (New) The method according to claim 146, wherein the mitochondrial disorder results in lower than normal uridine levels.

155. (New) The method according to claim 146, wherein the mitochondrial disorder is the result of prior or concurrent administration of a pharmaceutical agent.

156. (New) The method according to claim 155, wherein the pharmaceutical agent is a reverse transcriptase inhibitor, a protease inhibitor or an inhibitor of DHOD.

157. (New) The method according to claim 156, wherein the DHOD inhibitor is Leflunomide or Brequinar.

158. (New) The method according to claim 146, further comprising the administration of one or more co-factors, vitamins, or mixtures of two or more thereof.

159. (New) The method according to claim 158, wherein the co-factor is one or both of Coenzyme Q10 or calcium or magnesium pyruvate.

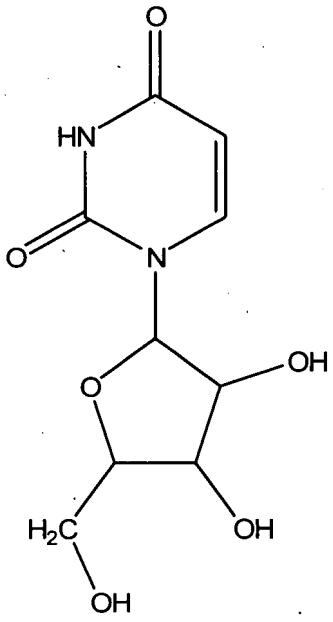
160. (New) The method according to claim 158, wherein the vitamin is selected from the group consisting of thiamine (B1), riboflavin (B2), niacin (B3), pyridoxine (B6), folate, cyanocobalamine (B12), biotin, α-lipoic acid, and pantothenic acid.

161. (New) The method according to claim 146, wherein the compound of Formula (I) or Formula (IA) is administered in a daily dosage in the range of about 0.5 g/m^2 to 20 g/m^2.

162. (New) The method according to claim 146, wherein the compound of Formula (I) or Formula (IA) is administered in a daily dosage in the range of about 2 g/m^2 to 10 g/m^2.

163. (New) The method according to claim 146, wherein the compound of Formula (I) or Formula (IA) is administered in a daily dosage of about 6.0 g/m^2.

164. (New) A method for reducing or eliminating one or more symptoms associated with a mitochondrial disorder comprising administering to a subject in need thereof an effective amount of L isomer or D isomer of a keto tautomer or a enol tautomer of a compound, the keto tautomer having the Formula I, and the enol tautomer having the Formula IA:



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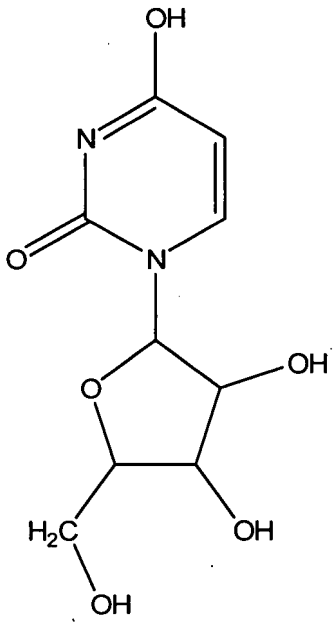

IA

wherein the mitochondrial disorder is selected from a group consisting of mitochondrial renal tubular acidosis, multiple mitochondrial deletion syndrome, Leigh syndrome, lactic acidemia, 3-hydroxybutyric acidemia, 1+proteinuria, pyruvate dehydrogenase deficiency, complex I deficiency, complex IV deficiency, aminoaciduria, hydroxyprolinuria, and MARIAHS syndrome, and wherein the compound is selected from uridine and 1-β-D-ribofuranosyluracil.

165. (New) The method according to claim 146, wherein the mitochondrial disorder is MARIAHS syndrome.

166. (New) The method according to claim 165, wherein the mitochondrial disorder comprises a deficiency in a pyrimidine synthetic pathway.

167. (New) The method according to claim 166, wherein the deficiency in a pyrimidine synthetic pathway is the deficiency in the uridine synthetic pathway.

168. (New) The method according to claim 166, wherein the deficiency comprises reduced expression and/or activity of an enzyme in the pyrimidine synthetic pathway.

169. (New) The method according to claim 168, wherein the enzyme is selected from the group consisting of dihydroorotate dehydrogenase (DHOD) and uridine monophosphate synthetase (UMPS).

170. (New) The method according to claim 165, wherein the mitochondrial disorder results in lower than normal uridine levels.

171. (New) The method according to claim 165, wherein the mitochondrial disorder is the result of prior or concurrent administration of a pharmaceutical agent.

172. (New) The method according to claim 171, wherein the pharmaceutical agent is a reverse transcriptase inhibitor, a protease inhibitor or an inhibitor of DHOD.

173. (New) The method according to claim 172, wherein the DHOD inhibitor is Leflunomide or Brequinar.

174. (New) The method according to claim 165, further comprising the administration of one or more co-factors, vitamins, or mixtures of two or more thereof.

175. (New) The method according to claim 174, wherein the co-factor is one or both of Coenzyme Q10 or calcium or magnesium pyruvate.

176. (New) The method according to claim 174, wherein the vitamin is selected from the group consisting of thiamine (B1), riboflavin (B2), niacin (B3), pyridoxine (B6), folate, cyanocobalamine (B12), biotin, α-lipoic acid, and pantothenic acid.

177. (New) The method according to claim 165, wherein the compound of Formula (I) or Formula (IA) is administered in a daily dosage in the range of about 0.5 g/m^2 to 20 g/m^2.

178. (New) The method according to claim 165, wherein the compound of Formula (I) or Formula (IA) is administered in a daily dosage in the range of about 2 g/m^2 to 10 g/m^2.

179. (New) The method according to claim 165, wherein the compound of Formula (I) or Formula (IA) is administered in a daily dosage of about 6.0 g/m^2.

180. (New) The method according to claim 164, wherein the mitochondrial disorder is MARIAHS syndrome.